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DISORDERS OF THE RESPIRATION AND CIRCULATION IN SUBJECTS WITH DEFORMITIES OF THE THORAX*

"If any reader imagines . . . let him know; that I am scarce five Feet high: that my Back was bent in my Mother's Womb: and that in Person I resemble Esop, the Prince of Orange, Marshall Luxemburg, Lord Treasurer Salisbury, Scarron, and Mr. Pope: not to mention Thersites and Richard the Third . . .

"it is natural to imagine . . . that generally deformed Persons would not be healthy or long lived. But this is a Question best determined by the facts . . ."

From *Deformity*, an essay by William Hay, Esq.
London, R. and J. Dodsley, 1754, pp. 4, 20

It has been known for centuries that individuals with deformed chests are subject to disorders of the respiration and circulation. With the advent of clinical-pathological correlations, it became clear that these disorders assume significant clinical proportions in only a few types of thoracic deformity, particularly in severe kyphoscoliosis. Such studies also suggest that shortening and distortion of the vertebral column may constitute a *sine qua non* for cardiorespiratory embarrassment; indeed, thoracic deformities which spare the spine, such as chicken breast, funnel chest and thoracoplasty, rarely interfere with cardiac or respiratory function.

In the last two decades, physiological measurements[1-3] have supplemented the clinical and anatomical observations. This new approach has made it possible to probe more deeply into the nature of the cardiorespiratory disturbances, particularly into their pathogenetic mechanisms. In this report, the respiration and circulation in the two types of thoracic deformity generally identified with heart or lung disease, *i.e.*, kyphoscoliosis and pectus excavatum, will be considered. Recent observations and measurements made in this laboratory[4] on such patients, before and after treatment, are the bases for the interpretations and conclusions which follow.

KYPHOSCOLIOSIS

Since the time of Hippocrates, the hunchback has been considered heir to an illness characterized by dyspnea, cyanosis and premature death. It has also been recognized that this illness involves both the heart and lungs. Only in recent years, however, has it been possible to establish that lung disease antecedes and effects the heart disease, and to relate the distortion of the thorax to specific patterns of abnormality in pulmonary performance.

Etiology and Anatomy

It may be exceedingly difficult to pinpoint the etiology of a skeletal deformity which begins, often insidiously, in childhood, and then progresses slowly during adolescence and adulthood. Indeed, in the majority of instances, the etiology of kyphoscoliosis either remains completely unknown or is ascribable to faulty development; these constitute the "idiopathic" group. The remainder are generally related either to muscle weakness, tuberculous spondylitis or injury. Of the group with muscle weakness, poliomyelitis is the most common cause, and only rarely may diseases such as muscular dystrophy, amyotonia or syringomyelia be implicated.

The factors which influence the degree of deformity include: (1) the initiating mechanism; (2) the number of affected vertebrae; (3) the age of onset; (4) the age of stabilization; and (5) the degree of dwarfing of the thorax. For example, tuberculous spondylitis, because of its destructive effect on the vertebrae, may effect a more severe degree of deformity than does muscle weakness; its disastrous effects are even further exaggerated if the infection strikes during childhood or adoles-

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cence, since deformity is abetted by rapid growth.

For some unexplained reason, right-sided scoliosis predominates over left. In this type of deformity, the hump, composed of the spinal deformity plus the attached ribs, lies to the right of the midline. The contour of the lungs is adapted to that of the thoracic cage by appropriate compression and distention. Although under such circumstances torsion and stretching of the great vessels seem almost inevitable, actual obstruction to the flow of either air or blood has not been demonstrated.

Only rarely is "pure" kyphosis or "pure" scoliosis associated with heart or lung disease; more often, a combination of these deformities has evolved by the time cardiorespiratory embarrassment is manifest.

Anatomical observations[5,6] have afforded little insight into the pathogenesis of the respiratory and circulatory disorders. Their deficiency may be attributed to several factors: (1) failure to distinguish between the asymptomatic kyphoscoliotic subject who dies from causes unrelated to his deformity, and the severely deformed patient who dies of respiratory and/or cardiac failure; (2) use of ambiguous anatomical terms, such as "emphysema," for which there is no *single* physiological counterpart; and (3) failure to dissociate the contribution of associated valvular disease, or systemic hypertension, to the cardiomegaly.

On the other hand, anatomical observations have provided a foundation for the interpretation of physiological measurements. For example, in the severely deformed patient, in whom disturbances in the respiration and circulation have dominated the terminal phase of the clinical course, certain abnormalities are characteristically present: (1) dilatation and hypertrophy of the right heart, and (2) small, distorted lungs and tracheobronchial tree, without pathognomonic alterations of the pulmonary parenchyma. Equally important, from a physiological point of view, is the lack of evidence for a primary role of tracheobronchial disease or obstruction in the evolution of heart or lung disease.

Clinical Assessment

Breathlessness, fatigability and, to a lesser degree, palpitation are the predominant complaints referable to the circulatory or respiratory apparatus. These are particularly manifest during exertion. By way of contrast, cough and wheezing are conspicuously absent. With the development of heart failure, pain in the right upper abdominal quadrant may attend hepatic engorgement.

Physical signs of abnormal respiratory and/or circulatory function may be detected even in the asymptomatic kyphoscoliotic subject. Thus, an abdominal type of breathing is characteristic, and short, vertical excursions of the rib cage usually substitute for normal, expansile motion. The pattern of breathing is generally one of small tidal volumes and rapid respiratory rates. Examination of the chest may disclose distant breath sounds over the gibbus; examination of the heart may reveal functional systolic and, on rare occasion, diastolic murmurs despite the absence of organic valvular heart disease.

Such abnormalities are exaggerated in the severely deformed. Some of these patients, particularly those with arterial hypoxemia and hypercapnia, may display clouding of the sensorium. Clubbing of the fingers and toes is not infrequent. Cyanosis is generally inconspicuous prior to the advent of cor pulmonale. When unequivocally present, it is usually associated with overt right heart failure, *i.e.*, systemic venous engorgement, tachycardia, and peripheral edema; at this time, it presumably reflects a combination of arterial hypoxemia and diminished blood flow through the superficial vessels of the skin.

Fluoroscopic and radiographic examination of the chest may be difficult to interpret in view of the rotation and distortion of the lungs and heart. Indeed, considerable experience may be required to avoid an erroneous diagnosis of organic heart disease. On the other hand, radiological examination may constitute the only reliable device for the display of pneumonitis, for the assessment of the range and pattern of diaphragmatic motion and for the estimation of the size of the heart.

The electrocardiogram generally affords little diagnostic help prior to the advent of cor pulmonale; at that time a pattern of right bundle branch block or right ventricular enlargement may appear. P waves may be prominent but their significance is difficult to assess. Finally, attention has been called to the occurrence of paroxysmal supraventricular tachycardia in kyphoscoliotic subjects; whether this coincidence reflects chance or whether the arrhythmia is attributable to the deformity remains to be established.

Pathophysiology

The cardinal, physiological defect elicited by kyphoscoliosis is alveolar hypoventilation. This abnormal state represents a failure of the chest bellows to deliver an adequate supply of ambient air to the gas-exchanging surfaces of the lungs. This insufficiency is manifested by a combination of systemic arterial hypoxemia and hypercapnia.

Some insight into the mechanisms which cause alveolar hypoventilation in kyphoscoliotic subjects is provided by measurements of lung volumes and observations on the mechanical performance of the chest bellows. The lung volumes, particularly the vital capacity, may be exceedingly small, corresponding to the degree of thoracic deformity and dwarfing. In most cases, the maximum breathing capacity is considerably reduced, reflecting, in large part, the musculoskeletal imbalance of the deformed cage. Such observations indicate a state of ventilatory insufficiency, origi-

nating in the chest bellows rather than in the lungs.

The mechanics of breathing are manifestly abnormal in these subjects; the abnormal mechanical performance is associated with an increase, to abnormal levels, in the work and energy cost of breathing. Both the chest and the lungs contribute to the increase in work; analysis of the nature of the resistances to breathing suggest that the elastic resistances of the chest, and to a lesser degree of the lung, are primarily involved. The increase in elastic resistances are, in turn, attributable to the musculoskeletal deformity and to the reduction in total lung capacity.

It has been pointed out above that the breathing pattern of the kyphoscoliotic subject is characterized by small tidal volumes and rapid respiratory rates. This spontaneous use of small tidal volumes minimizes the work required to overcome the augmented elastic resistances to breathing.[7] However, this adaptation is lost during exercise, since slight increments in the depth of breathing are associated with inordinate increments in the work of breathing.

A costly by-product of breathing with low tidal volumes and high respiratory rates is the sacrifice of alveolar ventilation for the sake of dead space ventilation. It is the consequences of alveolar hypoventilation, *i.e.*, arterial hypoxemia and hypercapnia, which may aggravate or precipitate failure of the respiration and circulation in kyphoscoliosis.

While hypoxemia profoundly influences the circulation, hypercapnia exerts its dire effects on the regulation of ventilation. The interplay of these factors and their relation to other abnormalities induced by the thoracic deformity are schematically represented in Figure 1. The dreaded *circulatory* sequelae of this interplay are right heart strain and failure; the corresponding *respiratory* sequelae are narcosis and death.

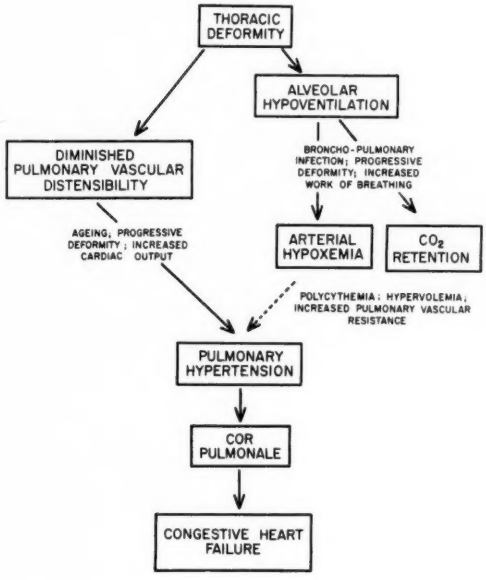

Figure 1. The evolution of cor pulmonale and CO_2 retention in kyphoscoliosis.

It is particularly interesting that despite the limitation imposed on the pulmonary vascular distensibility by the dwarfed and deformed thorax, only few circulatory abnormalities are manifest at rest. Indeed, prior to the advent of heart failure, the pulmonary artery and wedge pressures and the cardiac output are generally normal. During exercise, however, even though the oxyhemoglobin saturation of arterial blood may remain normal, an abnormal increment in pulmonary artery pressure consistently accompanies a normal increase in pulmonary blood flow; this pressor response is not attributable to back pressure from the left heart since pulmonary wedge pressures remain normal. In essence, the pulmonary pressor response to exercise presumably represents the effect of a normal increase in pulmonary blood flow and volume on a restricted vascular bed.

When in the course of the disease the sequelae, *i.e.*, polycythemia, hypervolemia, increase in pulmonary artery pressure and increase in cardiac output are superimposed, the circulatory burden may suffice to strain the right heart and ultimately to bring about its failure. At this time, pulmonary hypertension is present at rest and is associated with an abnormally low cardiac output; exercise fails to elicit the normal increment in pulmonary blood flow and exaggerates the pulmonary hypertension.

The effect of hypercapnia is quite different: in this circumstance, progressive accumulation of carbon dioxide is attended by loss of sensitivity of the respiratory center to the carbon dioxide, and possibly the pH, stimulus. A vicious cycle may then be established in which carbon dioxide retention, arising from inadequate alveolar ventilation, perpetuates itself by further depressing minute ventilation; if the rate of carbon dioxide accumulation is accelerated, as during acute pulmonary or bronchial infection, narcosis and death may ensue.

This combination of arterial hypoxemia and hypercapnia, and their physiological effects, resembles the advanced stages of chronic obstructive pulmonary emphysema. A common end result does not, however, imply a common pathogenesis. In chronic obstructive emphysema, the physiological defect arises from intrinsic lung disease and is characterized by impaired distribution of inspired air and mixed venous blood. In kyphoscoliosis, on the other hand, the predominant lesion arises from a defective chest bellows which, at least in its early stages, gives rise to

alveolar hypoventilation. It is only in the severely deformed and disabled kyphoscoliotic patient that the factor of inhomogeneity of ventilation and perfusion is superimposed on generalized alveolar underventilation.

The Management of Heart and Lung Disease in Kyphoscoliosis

It is clear that in kyphoscoliotic subjects, heart or lung disease may be prevented by minimizing the degree of deformity. In practice, it is difficult to translate this awareness into a definitive plan of orthopedic management since: (1) the application of orthopedic measures to the spine demands long-term immobilization and extensive surgical procedures; (2) these measures are most effective when applied to stabilize the spine in a young child or adolescent; and (3) it is difficult to estimate the degree to which childhood deformity may progress and elicit cardiorespiratory embarrassment in adulthood. Some aid in anticipating cardiorespiratory difficulty may be gained by repeated measurements of the vital capacity during the period of rapid development, since progressive reduction in this value presages the evolution of alveolar hypoventilation. Once alveolar hypoventilation and cor pulmonale are established, only medical management remains, since orthopedic correction is rarely feasible.

In the management of alveolar hypoventilation, the most dramatic results follow the use of mechanical respirators, even though the chest is severely deformed and apparently immobile. Their use improves alveolar ventilation, diminishes the work of breathing and relieves hypoxemia and hypercapnia. Adequate oxygenation of arterial blood may be expedited by substituting enriched oxygen mixtures for ambient air. Supplementary help in promoting the elimination of carbon dioxide may be afforded by the use of carbonic anhydrase inhibitors.[8] Such measures are of prime importance since relief of hypoxemia spares the heart and circulation, whereas the relief of hypercapnia not only avoids further depression of the respiratory center, but may possibly restore its sensitivity to normal chemical stimuli.

The use of pure oxygen or narcotics in patients with hypercapnia poses a distinct hazard. Both diminish ventilation, but each in a different manner: pure oxygen acts by abolishing the hypoxemic stimulus to ventilation, whereas narcotics further depress a center which is already partially refractory to normal chemical stimuli. Either of these measures may produce carbon dioxide narcosis and death unless a completely automatic respirator is substituted for the patient's own regulation of ventilation.

Once cor pulmonale exists, any further restriction of pulmonary function, as by bronchitis or pneumonia, may precipitate heart failure by aggravating systemic hypoxemia. At this time, the customary treatment for heart failure, digitalis, diuretics and salt restriction, is employed. However, the success or failure of treatment of this type of heart failure depends, in large part, on the adequacy with which hypoxemia and its sequelae are combated. Thus, the therapeutic program leans heavily on mechanical respiratory aids to augment alveolar ventilation, on antibiotics to clear bronchopulmonary infection, and on venesection to reduce the hemodynamic burden imposed by polycythemia. If a bronchial infection introduces a bronchospastic element, bronchodilators may promote patency of the smaller airways.

PECTUS EXCAVATUM (FUNNEL CHEST)

Anatomy and Etiology

Funnel chest is a thoracic deformity characterized by posterior dislocation of the body of the sternum and its attached ribs; the affected portion of the sternum is concavely deformed as well as displaced. The manubrium is generally normally situated, whereas the xiphoid may extend so far posteriorly as to impinge on either the vertebral bodies or the paravertebral gutters.

Although, in the case of mild deformity, the heart may retain its usual position between the sternum and vertebral column, in the more severe forms of sternal depression the heart is often dislocated into one hemithorax, generally the left. Despite the bony encroachment on the mediastinal space, and the displacement of the mediastinal contents into the area usually occupied solely by the lungs, the thoracic volume, as reflected in measurements of lung volumes, remains remarkably unaffected.

The nature of the defect is not well understood. A congenital basis is suggested by its frequent recognition at, or shortly after, birth, and by its occurrence in patients with syndromes, such as Marfan's, which include widespread congenital defects. An hereditary influence is indicated by its appearance in multiple members of a single family.

The mechanical basis for the defect also remains to be established; proposed mechanisms include overgrowth of ribs, shortening or contracture of the central tendon of the diaphragm and infantile respiratory obstruction.[9,10]

Symptoms and Signs

As a rule, the patient is asymptomatic at rest. During exertion, breathlessness, palpitation and easy fatigability are common complaints. A particular problem in assessing such complaints is the psychological disturbance which attends skeletal deformity, particularly in older children and adolescents.

Objective evidence for cardiorespiratory involvement is usually lacking. Systolic murmurs may be heard over the precordium; diastolic murmurs require a search for independent valvu-

lar disease. Overt failure of either the right or left heart is rarely attributable to the deformity *per se*. Heart failure seems most apt to occur when the deformity is complicated by some other independent cardiac disorder such as atrial fibrillation or systemic hypertension.

Fluoroscopic and radiographic examination may be misleading if it is not recognized that broadening of the transverse diameter of the heart may merely reflect anatomical adaptation of a normal heart to the narrowed anteroposterior diameter of the chest. The electrocardiogram is generally normal.

Physiological Considerations

There are few measurements of physiological variables, other than lung volumes, in subjects with pectus excavatum. As indicated above, they are generally normal or near-normal, even in those who are severely symptomatic. Other ventilatory measurements, such as estimates of the maximum voluntary ventilatory capacity, arterial blood gaseous composition, and alveolar-capillary gas exchange, are also within normal limits. Observations on the circulation, both at rest and during exercise, have revealed normal pulmonary blood flows and pulmonary artery pressures.

It is to be emphasized that these comments apply to patients who are symptomatic but who present no objective evidence for heart failure. On the other hand, Ravitch[11] has recorded physiological measurements in a young man with heart failure. This patient also had atrial fibrillation. In this subject, surgical correction restored an elevated pulmonary artery pressure and a severely curtailed cardiac output to normal levels; end-diastolic blood pressures in the right ventricle were normal pre- and post-operatively. More observations of this type, particularly in instances of heart failure which are not complicated by unrelated cardiac disorders, should help to define the nature of the cardiac overload in pectus excavatum.

Treatment

If it can be established that the heart has been compromised by the deformity, it would seem reasonable to release it surgically from its abnormal position. However, it is only on rare occasions that objective evidence for cardiac embarrassment has been adduced. Even in these instances the mechanism responsible for the cardiac failure has not been clarified. Since surgical intervention for cosmetic, psychological and cardiac relief promises to continue, and since the appropriate tools for physiological investigation in man are available, considerable insight may be expected into the nature and degree of the cardiac disorder which may be imposed by pectus excavatum.

SUMMARY

Two types of thoracic deformity, *i.e.*, kyphoscoliosis and pectus excavatum, have been singled out for review with respect to their effects on the respiration and circulation. They stand in striking contrast: kyphoscoliosis emerges as a mechanism for initiating a train of events which may terminate in cor pulmonale and heart failure; pectus excavatum, on the other hand, appears to inflict predominantly cosmetic and psychogenic injury and rarely, if ever, compromises cardiac function.

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June 13th deadline . . .

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FOR ABSTRACTS OF PAPERS
31ST ANNUAL SCIENTIFIC SESSIONS
AMERICAN HEART ASSOCIATION

Abstracts for the 31st Annual Scientific Sessions of the American Heart Association, to be held in San Francisco, October 24-26, 1958, will be accepted on special forms which can be obtained from Dr. F. J. Lewy, American Heart Association, 44 East 23rd Street, New York 10, New York.

Abstracts must not exceed 250 words and should contain, in summary form, results obtained and conclusions reached. They should be submitted in duplicate and must be suitable for publication in the Proceedings without further editing.

The deadline is June 13, 1958. Abstracts which arrive late, or are longer than 250 words, cannot be considered.

Space for Scientific Exhibits may be requested on application forms also available at the address above. ***The deadline for these applications is June 13, 1958.*** They will be submitted for decision to the Committee on Scientific Sessions Program of the Association.

ANNUAL MEETING OF THE AMERICAN HEART ASSOCIATION

and

THE 31ST SCIENTIFIC SESSIONS

October 24-28, 1958 **San Francisco, California**

THIRD WORLD CONGRESS OF CARDIOLOGY

Brussels **September 14-21, 1958**

IMPORTANT REMINDER!

The officers of the Third World Congress of Cardiology wish to remind all physicians who plan to attend the Congress that registration blanks and fees should be forwarded as soon as possible.

The deadline for registration is set irrevocably for May 1, 1958.

Information about the Congress, and blanks for registration and hotel accommodation, may be obtained from the Inquiry Section, American Heart Association, 44 East 23rd Street, New York 10, New York.

For special information (other than registration and blanks) write directly to: Dr. Fr. van Dooren, Secretary of the Congress—80, rue Mercelis, Brussels, Belgium.

For travel arrangements and information on pre- and post-Congress tours, write or telephone: Convoys Travel Service, 1133 Broadway, New York 10, New York. Telephone: WAtkins 9-0920. *Special economy fares are available.*